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Case Report

Unilateral Direct High-Flow Traumatic Carotid-Cavernous Fistula Presenting as Isolated Bilateral Abducent Nerves Palsy: A False Localizing Sign

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Abstract

Direct CCF most commonly occurs as a result of trauma, forming a direct fistula between the internal carotid artery and the cavernous sinus. As the high pressure of the internal carotid artery is transmitted to the cavernous sinus, it commonly leads to oculomotor, trochlear and abducens cranial nerve palsy. The case report described herein describes young adult presented with recent onset right eye proptosis and binocular horizontal diplopia due to bilateral lateral gaze palsy. The patient had history of RTA 1 year back. The patient was initially treated for facial bone complex fractures, following which due to persistent symptoms she was then evaluated for infectious and neoplastic etiology. The patient was diagnosed with right CCF on CTA. Endovascular embolization was done resulting in resolution of proptosis and improved prognosis. However, It is very rare for post-traumatic, unilateral CCF to present with contralateral symptoms and bilateral abducent palsy without affecting other cranial nerved in direct CCF. Therefore, patients presenting with B/L abducent palsy should evaluated for CCF with either a CTA/DSA as early diagnosis and treatment results in improved patient outcome.

Keywords: RTA: Road Traffic Accident; CCF: Carotid- Cavernous Fistula; CTA: Commuted Tomography Angiography; DSA: Digital Substraction Angiography; ORIF: Open Reduction and Internal Fixation

Introduction

CCFs are arteriovenous malformations that result in shunting of the blood from the carotid artery to the cavernous sinus. Pulsating exophthalmos, bruit, and conjunctival chemosis have long been regarded as the three classical symptoms of carotid cavernous fistula (CCF). However, the clinical presentation depends upon the specific venous drainage of the cavernous sinus. CCFs can be classified by Barrow classification [1]. according to the lesion that connects the cavernous sinus to the carotid artery. Type A CCFs have a direct high flow connection between the internal carotid artery and the sinus. Type B CCFs are dural shunts between the meningeal branches of the internal carotid artery and the sinus. Type C form from dural branches of the external carotid and type D involve meningeal branches from both the external and internal carotid and the cavernous sinus. Direct CCF are known to cause combined 3^{rd},4^{th} and 6^{th} nerves palsy due to increased pressure inside cavernous sinus, however it is very rare for direct high flow fistula's to present with isolated Bilateral 6^{th} nerve palsy without involving the 3^{rd} and 4^{th} nerve. Therefore, the possible pathophysiology and mechanism needs to be explored. Also,

patients having craniofacial trauma can have similar presentation, hence a high suspicion of CCF should be maintained in patients having such clinical presentation.

Case Presentation

A young adult patient presented with history of proptosis of right eye along with binocular horizontal diplopia for 1 year now. The patient had history of trauma due to RTA 1 year back for which ORIF of the right zygoma and mandible was done. Following the surgery, the symptoms persisted.

On clinical examination, the patient kept the head rotated to the left side at rest. The patient had non-axial proptosis of the right eye with conjunctival congestion and dilated episcleral vessels.

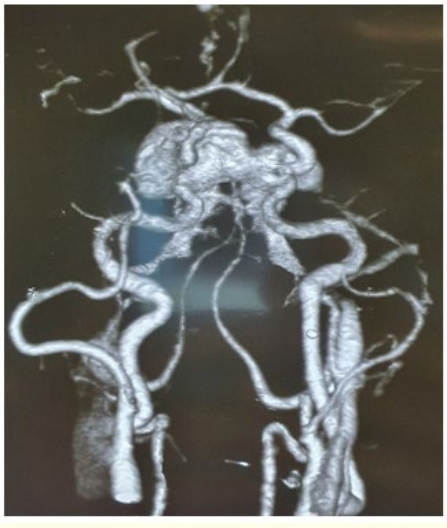

Figure 1

Both eyes had systolic bruit on auscultation, far vision was 6/60 in the right eye and 6/9 in the left eye. Also had restriction of movement on abduction suggestive of bilateral abducens palsy, affecting the left eye more than the right eye. The diplopia was binocular and present at all positions of gaze, increased on looking at distant objects and decreased on looking at near objects. The patient was evaluated for possible diagnosis of CCF was suspected.

CTA was done which was suggestive of bulky Rt. Cavernous sinus with dilated superior and inferior ophthalmic veins. DSA was done showed a direct, high flow fistula between the cavernous ICA and the right cavernous sinus with retrograde filling of the superior and inferior ophthalmic veins as well as the opposite cavernous sinus and B/L inferior petrosal sinuses along with the basilar plexus. No cortical/deep venous reflux was seen.

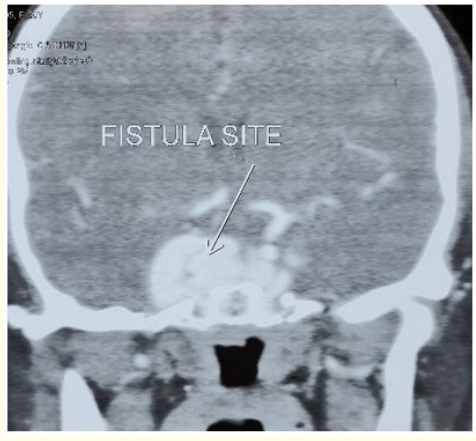

Figure 2

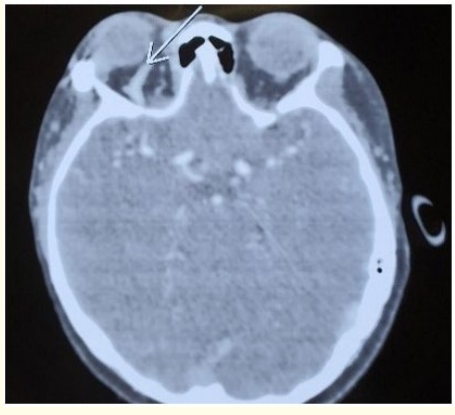

Figure 3

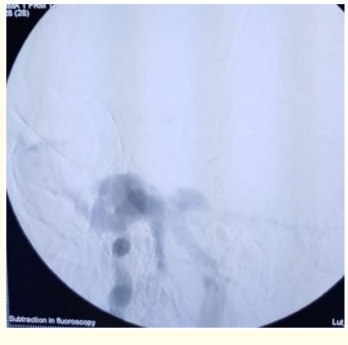

Figure 4

Figure 2,3,4: CT angiography is suggestive of dilated B/L cavernous sinuses [Right>left] with dilated Rt. Superior and inferior ophthalmic veins.

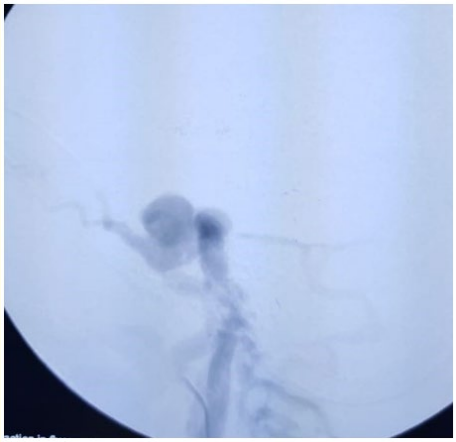

Figure 5

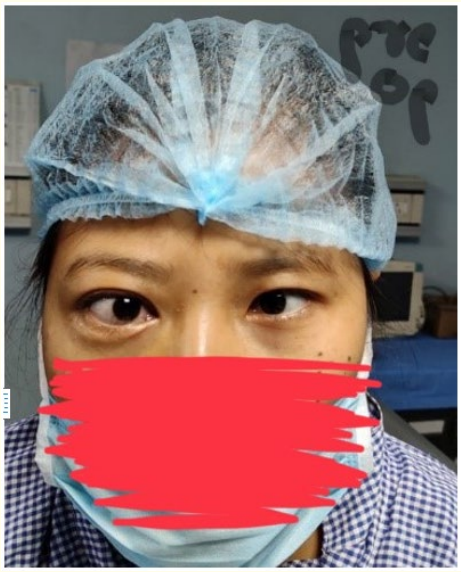

Figure 6

Figure 5,6: Catheter angiography showed a high flow fistula between the cavernous Rt. ICA and Rt. Cavernous sinus with anterior and posterior drainage into the ophthalmic veins and B/L Inferior petrosal sinuses, with no cortical drainage/reflux.

The differential diagnosis for bilateral abducens nerve palsy is broad, but if diplopia is of sudden onset, vascular aetiologies such as aneurysms with subarachnoid haemorrhage, vascular malformations, or ischemic stroke need to be considered. Other potential causes of bilateral abducens nerve palsy include (1) infective or infiltrative diseases such tuberculosis and syphilis; (2) demyelinating diseases such as multiple sclerosis; (3) increased intracranial pressure due to space-occupying lesions or idiopathic intracranial hypertension, which can affect the abducens nerve as it transverses in the prepontine cistern; (4) pathology in the cavernous sinus such as cavernous ICA aneurysm, CCF, infection that can affect the abducens nerves as they course through the cavernous sinus (since both cavernous sinuses communicate with one another); and (5) brainstem lesions (6) or in some rare instances, thyroid eye disease [2]. Orbital lesions or myasthenia gravis should also be considered in these cases. There were no signs of raised intracranial pressure. MRI of the brain and orbits did not show any signs of compression along the course of the sixth cranial nerves or brainstem pathology. The CT angiogram and catheter angiogram confirmed the CCFs.

Coil embolization was done via trans-arterial route, resulting in significant improvement in the symptoms.

Discussion

Our patient had a direct high-flow CCF that most likely occurred due to trauma. Clinical symptoms and signs of CCF depend on a variety of factors, including fistula size, location in the cavernous sinus, drainage route (anterior, posterior, or both) and rate of blood flow [3,4]. Therefore, the Barrow classification is not very practical from clinical and therapeutic standpoints, as symptomatology and current treatment approach are influenced largely by venous drainage. Today, the venous drainage of CCFs is of greater importance for the management of these lesions and has to be a key component of any contemporary classification system. Thomas., *et al.* proposed a classification system using venous drainage that captures the symptomatology, endovascular treatment approach, and outcome [5].

CCFs are a rare cause of bilateral abducens nerve palsy and may occur spontaneously or in the context of trauma. Since the sixth nerve is in close proximity to the internal carotid artery in the cavernous sinus, it is vulnerable when CCFs develop. The mechanism is thought to involve direct compression or vascular steal with cranial nerve ischaemia [6,7]. Another potential mechanism of limited eye movements is venous congestion resulting in swelling of the extra-ocular muscles, but there was obvious difference in the size of the lateral rectus muscle on imaging in this case, making it a likely possibility, but it doesn't explain the occurrence of lateral rectus palsy in the opposite eye.

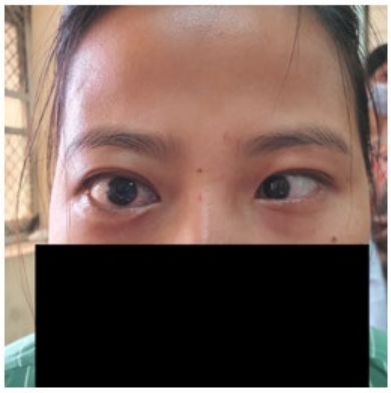

Figure 7: Eye position at rest, which is suggestive of B/L abducens palsy.

However, these mechanisms do not explain the cause of isolated bilateral abducens CN palsy without affecting oculomotor or the trochlear nerves.

Relevant ANATOMY OF THE ABDUCENS NERVE: Dorello's canal was described for the first time, by Gruber, in 1859 AD, as an osteofibrous canal at the apex of the petrous bone, containing the abducens nerve and the inferior petrosal sinus. Dorello's canal may be simplified, as a canal, between a point below the posterior clinoid process and the most anteromedial portion of the petrous ridge. It is 4.0 to 13 mm in length, and from 0.5 to 3.0 mm in diameter. he posteromedial limit of the canal is formed by a ligament, connecting the petrous ridge to the clivus (the petrosphenoidal ligament or Gruber's ligament). The lateral wall is formed by the anteromedial sphenoid ridge and the medial wall by the clivus. The whole canal is embedded inside a venous confluence which occupies the space between the two dural layers of the petroclival area.

The dural entrance porus of the abducens nerve was surrounded by the IPS [8]. Therefore, venous hypertension in the IPS causes compression of the abducens nerve and thereby abducens palsy. In our case also, Bilateral IPS sinuses are dilated, and this probably has caused Bilateral abducens palsy in our patient without affecting other cranial nerves.

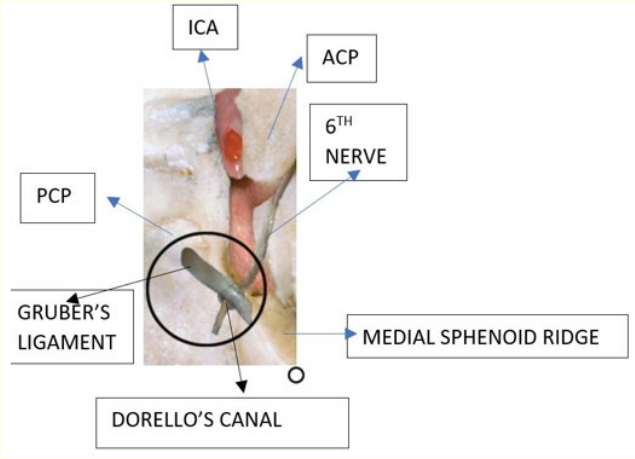

Figure 8

When these symptoms and signs are present in a patient with a history of craniomaxillofacial trauma, post-traumatic CCF must be suspected. In the case presented, diagnosis of post-traumatic carotid aneurysm initially and CCF later was difficult for two reasons. The first reason was the associated facial fractures of the zygomatic complex, nasal bones, and orbital floor which cause similar findings and led to the first false diagnosis of post-traumatic superior orbital fissure syndrome. Second, the osteosynthetic and alloplastic material that were used around the left eye led to the other false diagnosis of postoperative infection/orbital cellulitis. The persistent clinical signs led to the right diagnosis of CCF. However, a high index of suspicion and a deep knowledge of this pathologic entity would probably lead to the right diagnosis from the beginning [9,10]. In conclusion, CCFs should be considered in the differential diagnosis of bilateral abducens nerve palsy. A detailed examination to assess for signs of this such as dilated episcleral vessels should be carried out in the context.

Conclusion

- When these symptoms and signs, such as ophthalmoplegia/ ocular cranial nerves palsy is present in a patient with a history of craniomaxillofacial trauma, post-traumatic CCF must be suspected.
- Computed tomography angiography and magnetic resonance angiogram) though are useful non-invasive imaging modalities, and catheter angiography/DSA remains the gold

standard, and is a must for all cases to confirm the diagnosis and for the operative planning. It also re-affirms that a better venous drainage- based classification is needed which better explains the symptomatology, therapeutic approach as well as the outcome.

- Patients should understand that the abducens nerve palsy may worsen in spite after treatment, although there have been a good rate of reported spontaneous improvement [11].

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Conflict of Interest

None.

Bibliography

1. Barrow DL., *et al.* "Classification and Treatment of Spontaneous Carotid-Cavernous Sinus Fistulas". *Journal of Neurosurgery* 62.2 (1985): 248-256.
2. Keane James R. "Bilateral Sixth Nerve Palsy". *Archives of Neurology* 33.10 (1976): 681.
3. Ellis Jason A., *et al.* "Carotid-Cavernous Fistulas". *Neurosurgical Focus* 32.5 (2012): E9.
4. Robert Thomas., *et al.* "Thrombosis of Venous Outflows of the Cavernous Sinus: Possible Aetiology of the Cortical Venous Reflux in Case of Indirect Carotid-Cavernous Fistulas". *Acta Neurochirurgica* 159.5 (2017): 835-843.
5. Thomas Ajith J., *et al.* "Proposal of Venous Drainage-Based Classification System for Carotid Cavernous Fistulae with Validity Assessment in a Multicenter Cohort". *Neurosurgery* 77.3 (2015): 380-385.
6. Sivaruben Kalaichandran and Jonathan A Micieli. "Bilateral Sixth Nerve Palsies from Carotid Cavernous Fistulas with Transient Worsening Following Transvenous Embolisation". *BMJ Case Reports* 12.11 (2019): e232191-e232191.
7. Zhu Linxin., *et al.* "Post-Traumatic Right Carotid-Cavernous Fistula Resulting in Symptoms in the Contralateral Eye: A Case Report and Literature Review". *BMC Ophthalmology* 18.1 (2018).
8. Ezer Haim., *et al.* "Dorello's Canal for Laymen: A Lego-like Presentation". *Journal of Neurological Surgery Part B: Skull Base* 73.03 (2012): 183-189.
9. Lazaridou Maria., *et al.* "Carotid-Cavernous Fistula as a Complication of Facial Trauma: A Case Report". *Craniomaxillofacial Trauma and Reconstruction* 8.3 (2015): 239-245.
10. Peng Teng J., *et al.* "Carotid-Cavernous Fistula Presenting with Bilateral Abducens Palsy" 51.6 (2023).
11. Zanaty Mario., *et al.* "Endovascular Treatment of Carotid-Cavernous Fistulas". *Neurosurgery Clinics of North America* 25.3 (2014): 551-563.